ECTOPIA CORDIS‡

(A case report and review of literature)

by

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"Ectopia Cordis" is a condition in which the heart is situated in an abnormal position. It was first described by Haller (Blatt and Zeldes) in 1706. It has been classified by Townsend in 1833 (Logan) as follows:—

1. Cervical — having an intact sternum with heart in the cervical region.
2. Thoraco-cervical — there is defect in the cranial end of the sternum with the heart partially in the cervical region.
3. Pectoral heart with fissure of the sternum — the sternum presents various deficiencies and the heart lies outside the thorax.
4. Abdominal heart — with the defect in the diaphragm.
5. Thoracico-abdominal.

The case reported below, which occurred at Nowrosjee Wadia Maternity Hospital, had a pectoral heart associated with many other malformations of viscera. The case is reported for its rarity.

Case Report:

A patient, aged 30 years, was admitted on 3rd December 1966 with a history of 7 months' amenorrhoea and pain. She had slight bleeding per vaginam for one day. The patient had visited the antenatal department only once on 19th November 1966 and no unusual finding was detected.

She had been married for 12 years. She was the second wife. The husband had a 10 year old normal child by his first wife who is alive. The patient had an abortion of 4 months and another at 3 months, six years and nine years respectively after her marriage. There was no history of venereal disease in the couple. There was no history of taking any drug or of jaundice or infective fevers during this pregnancy.

On 3rd December, the patient, when she was 30 weeks pregnant, delivered a macerated foetus immediately on admission. The third stage of labour was uneventful.

On examination of the foetus, weighing 400 gms., the following abnormalities were noted:

It was a female with the heart covered with the pericardium lying outside the thoracic cavity anteriorly as shown in the photograph. There was no other malformation externally, but dissection revealed many other malformations associated with the pectoral heart.

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The part of the sternum below the manubrium was absent. The left dome of the diaphragm was absent while the right dome of the diaphragm was normally developed. The left lung was 1/4th the size of the right lung. The remaining 'left side of the thoracic cavity was occupied by parts of small intestines, large intestines, stomach and spleen.

The caecum was lying almost in the epigastric region. The kidneys were horse-shoe shaped, with the connecting bar at the level of the pelvic brim. The ureters were single on each side. Further dissection of the heart did not reveal any abnormalities. The great vessels of the heart were normal.

The following investigations in the mother were carried out:

Hb. 12 gms.%; blood group A Rh positive, V.D.R.L. negative; urine normal, blood urea—14.4 mg.% (higher side of normal); glucose tolerance curve normal; Listeriosis—smear and culture both negative. No definite cause for the malformations could be accounted for.

Comments and Review of literature

The heart and diaphragm arise in the cervical region and descend during the fifth to the tenth weeks of gestation. According to Logan (1965), the defects leading to "ectopia cordis" are apparent by the sixth to seventh week of gestation and are considered inherent abnormalities of germ plasm bearing no relationship to parenteral health and to complications of pregnancy or delivery. In the case described by Greiffenberg (quoted by Birnhaum), the amnion had acquired an adhesion to the heart and interferred with the closure of the thoracic cavity. As a result of the drag produced by the amniotic adhesion, the heart was subsequently drawn out of the closing thorax. The rarity of the condition may be explained by the comparatively short time during which the heart is exposed to such influences.

Up till now 54 cases of ectopia cordis have been described in literature. Out of these, major group was that of the thoracic variety. In most instances delivery was normal. In Welch's case (1909) (Blatt and Zeldes), the heart ruptured during delivery.

The condition is usually not diagnosed till the baby is delivered, but an interesting case has been described by Lumsden, in 1960, where he had succeeded in diagnosing ectopia cordis clinically in utero during labour. The case history in brief was as follows:

An elderly gravida 2, aged 40 years, had vaginal bleeding in early pregnancy and subacute hydramnios, beginning at 29th week of gestation. X-ray prior to labour showed a single foetus without any evidence of abnormality. She was admitted at 37th week for high rupture of membranes, 30 ounces of yellow coloured liquor were drained. Contractions began 2 hours later. Labour was carefully watched and it was noted that even with moderate contractions, the foetal heart sounds slowed to 10(beats per minute, recovering rapidly to 140 per minute in the intervals. With strong contractions the heart could not be heard but returning strongly as the contractions died away. The foetal position was difficult to define at first but as the liquor drained away palpation became easy. During the latter part of labour, the foetal back was directly anterior, both shoulders being easily palpated and yet the foetal heart sounds could no be heard anywhere over the back.

On the other hand, the sounds were heard strongly far out in each flank. Taken in conjunction with the disappearance of the heart sounds during contractions and their rapid reappearance in the intervals, this phenomenon could be explained by the heart lying outside the body wall and a diagnosis of ectopia cordis was made. A female child weighing 7 lbs. was delivered by low forceps.

The baby had eventration of heart, liver, spleen and intestines. The heart was beating strongly on delivery and peristaltic movements of the gut were active. No attempt was made to resuscitate the child because of gross deformities incompatible with life. The child did not make any effort to breathe. In spite of this the heart continued to beat strongly for approximately twelve hours and it is suggested that sufficient oxygen diffused through the peritoneal and pericardial coverings of the ectopic viscera to supply the child's requirements.

The delivery may be premature as in many of the reported cases, or in a few at term. Duration of life depends on the extent of malformation. A few of the cases reported were stillborn and many died within few hours of birth. The duration of life was thirty-six hours on an average, while patients with only minor defects have been diagnosed for the first time during adult life. Of the types of ectopia cordis described, cervical one appears to be incompatible with life. The thoracic offers a better prognosis depending on extent of malformation, while with the abdominal type a number of patients have lived well into adult life, sometimes undiagnosed till then (Blatt and Zeldes, 1942).

When gross malformation is present, though the life expectancy is very short, an attempt should always be made to provide a protection and coverage to the exposed heart. This is the main surgical consideration, although in many cases the sutures had to be released immediately because of rapid embarrassment of the heart action. A few patients have been cured when there has been a very minor defect.

Summary

An interesting case of ectopia cordis is reported with a short resume of the subject and literature.

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